

SECTION 23

Endocrinology

USAMA ALJUMAILY

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1. The hypothalamus secretes releasing or inhibiting factors to control the anterior pituitary gland.

Of the following, the pituitary hormone that is elevated in a hypothalamic disease is

- A. thyroid-stimulating hormone
- B. follicle-stimulating hormone
- C. luteinizing hormone
- D. prolactin**
- E. adrenocorticotrophic hormone

Prolactin is the only pituitary hormone that is suppressed by a hypothalamic factor, dopamine. Hypothalamic deficiency leads to a decrease in most pituitary hormone secretions but may lead to an increase in prolactin secretion.

2. You are evaluating a newborn baby with cleft palate; his mother asks you about any risk of associated congenital hypopituitarism.

Of the following, this baby is **MOST** likely at risk for

- A. hypothyroidism
- B. hypogonadism
- C. hypoprolactinemia
- D. anti-diuretic hormone deficiency
- E. growth hormone deficiency**

Hypopituitarism

Clinical picture 1. At birth: - Normal size (near normal weight and height) - Micropenia is a diagnostic clue: - May be neonatal emergency as apnea, cyanosis, hypoglycemia - May be mild facial anomalies e.g. cleft lip & palate	
2. Later on: Severe growth failure - Proportionate short stature - Height below the 1st percentile for age and sex - or height < 2 SD below sex-adjusted Mid-parent height - Growth velocity < 5cm/year — Arrested by the end of the 2nd year	 Childhood features - Small face, nose & mandible - Prominent forehead & depressed nasal bridge - Wide anterior fontanel & late bone - Delayed teething
- Micropenia in childhood; normal for body size in adults - Hypopigmentation in infants and children; fasting symptoms in some adults	

3. During a routine clinical visit, a healthy asymptomatic 6-year-old boy is found to have fasting blood glucose of ^{>95}110 mg/dL; he has normal oral glucose tolerance test, no glycosuria, and HbA1c of 6.4%. *normal.*

This child is **MOST** likely considered to have

- A. impaired fasting glucose**
- B. impaired glucose tolerance
- C. sporadic hyperglycemia
- D. type 1 diabetes mellitus
- E. normal glucose metabolism

4. In insulin-dependent diabetes mellitus, hyperglycemia results when insulin secretion becomes inadequate to enhance peripheral glucose uptake and to suppress hepatic and renal glucose production.

Of the following, the sign of more complete insulin deficiency is



- A. postprandial hyperglycemia
- B. fasting hyperglycemia
- C. ketogenesis
- D. glycosuria
- E. metabolic acidosis

5. A 5-year-old boy develops vomiting and abdominal pain with signs of dehydration. He has been diagnosed with type 1 diabetes mellitus since the age of 4 year and he is on insulin therapy since that time.

Of the following, the sign that differentiate this condition from gastroenteritis is

- A. polyuria
- B. polydipsia
- C. abdominal tenderness
- D. tachypnea
- E. altered mental status

The presence of polyuria, despite a state of clinical dehydration, indicates osmotic diuresis and differentiates patients with DKA from patients with gastroenteritis or other gastrointestinal disorders.

6. In DKA, electrolyte abnormalities occurs secondary to hyperglycemia and acidosis. Which of the following electrolytes is artificially measured low in DKA?

- A. potassium
- B. sodium
- C. phosphorus
- D. calcium
- E. bicarbonate

The measured serum sodium concentration is artificially low, however, because of hyperglycemia. Hyperlipidemia also contributes to the decrease in measured serum sodium.

7. A 9-year-old girl develops polyuria, vomiting, abdominal pain, and deep acidotic breathing during an intercurrent illness. She has been diagnosed with type 1 diabetes mellitus 5 years ago. Lab investigations reveal: blood glucose, 600 mg/dL; blood pH, 7.28; serum bicarbonate concentration, 14 mEq/L; and ketones in urine are positive.

Of the following, the **MOST** serious complication that may occur in this girl is

- A. cardiac arrhythmias
- B. cerebral edema
- C. acute tubular necrosis
- D. pulmonary edema
- E. bowel ischemia

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بثاني ساعه انطى 0.45

ومائطي بايكاربونات ابد

8. You are going to treat a 7-year-old boy with type 1 diabetes mellitus who develops DKA due to a recent infection. Examination reveals severe dehydration

and deep acidotic breathing. Lab investigations shows: blood glucose, 450 mg/dL; blood pH, 7.2; and serum bicarbonate concentration, 14 mEq/L.

Which of the following should be avoided during the treatment of this girl?

- A. lactated Ringer solution
- B. glucose
- C. bicarbonate
- D. fast-acting soluble insulin
- E. potassium phosphate

Bicarbonate therapy should be avoided as it increases risk of the development of cerebral edema.

9. Measurement of hemoglobin A1c (HgbA1c) provides a mean for assessing long-term glycemic control for children with diabetes mellitus. In which of the following conditions this measurement is inaccurate for long-term glycemic control?

- A. children with hypoalbuminemia
- B. children with cystic fibrosis
- C. children with malabsorption
- D. children with hemoglobinopathies
- E. children with chronic liver disease

10. A 5-year-old girl has been developed type 1 diabetes mellitus 6 months ago. She is on intensive control using multiple daily injections of insulin. A recent blood testing of HgbA1c is 6.5%.

Of the following, the **MOST** frequent complication that may be encountered at this age is

- A. recurrent hypoglycemia
- B. recurrent hyperglycemia
- C. weight loss
- D. genital candidiasis
- E. recurrent urinary tract infections

11. Type 2 DM can be managed with lifestyle modifications, including nutrition therapy, increased exercise, and oral hypoglycemic agent (metformin); however, insulin therapy is necessary in special circumstances.

Of the following, insulin treatment is required when there is

- A. fasting hyperglycemia
- B. postprandial hyperglycemia
- C. hyperkalemia
- D. hypophosphatemia
- E. ketonuria

Like DKA

12. Hypoglycemia may be a manifestation of child abuse or Munchausen syndrome by proxy induced by exogenous administration of insulin (i.e., factitious hyperinsulinemia).

Of the following, the **MOST** reliable test that confirms the diagnosis of factitious hyperinsulinemia is

- A. very low concentration of C-peptide
- B. very low level of blood glucose
- C. presence of ketones in urine
- D. presence of ketones in serum
- E. high level of insulin concentration

This diagnosis should be suspected if extremely high insulin concentrations are detected ($>100\mu\text{U/mL}$). C-peptide concentrations are low or undetectable, which confirms that the insulin is from an exogenous source.

13. Hypoglycemia may result from absence or deficiency of the counter-regulatory hormones (growth hormone and cortisol). It is sometimes difficult to differentiate it from hyperinsulinism.

Which of the following suggests hypoglycemia due to absence of counter-regulatory hormones rather than hyperinsulinism?

- A. presence of serum ketones
- B. very low level of blood glucose
- C. absence of ketones in urine
- D. high level of GH and cortisol
- E. high level of insulin concentration

فاتطي انسولين

hypoglycemia
 ↗ factitious hyperinsulinemia → ↓ C-peptide
 ↘ non-hyperinsulinemia → Ketonuria

14. The upper-to-lower segment ratio is one of methods for growth assessment that changes with age.

Of the following, this ratio becomes 1:1 at the age of

- A. two year
- B. four year
- C. six year
- D. eight year
- E. ten year

15. A 12-year-old male adolescent is found to have short stature; lab tests and radiological study confirms idiopathic growth hormone (GH) deficiency. You decide to start treatment with biosynthetic recombinant DNA-derived GH.

This adolescent is especially at high risk for development of

- A. pseudotumor cerebri
- B. leukemia
- C. carpal tunnel syndrome
- D. diabetes mellitus

2. Recombinant growth hormone

"FDA approved indications of GH therapy":

- GH deficiency
- Turner syndrome
- Chronic renal failure before transplantation
- Idiopathic short stature
- Small-for-gestational age short stature
- Prader-Willi syndrome
- Noonan syndrome

High risk to develop slipped capital femoral epiphysis

E. slipped capital femoral epiphysis

16. You are discussing short stature (SS) with medical students. You mention that congenital growth hormone deficiency is one of common causes of SS. An important statement that should be included with your discussion is

- A. infants usually have a normal birth length and weight at term
- B. growth rate accelerates after birth till the age of 2 to 3 years, then declines
- C. these children have a normal weight-to-height ratio
- D. infants are usually thin and slim
- E. microphallus is usually a late presentation in male gender

17. The staging of pubertal changes differs between both genders. The onset of puberty is marked by pubarche and gonadarche.

Of the following, the first normal event of pubertal maturation in boys is

- A. enlargement of the testis
- B. appearance of the pubic hair
- C. thinning of the scrotum
- D. deepening of the voice
- E. appearance of the axillary hair

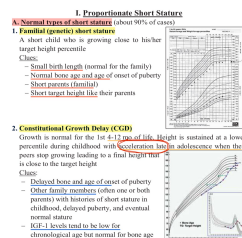
Puberty	
Definition - It is a period of growth lasting 5 years, consisting of 3 stages and includes physical, sexual & psychological changes. - Onset Girls: 8-13 years, Boys: 9-14 years - Sequence :	
Girls	Boys
- Breast development - Pubic hair - External genitalia maturation - Feminine habitus - Axillary hair - Oil secretion and acne - Menstruation	- Testicular growth - Pubic hair - Penis and scrotal growth - Increased muscle bulk - Body hair (beard, axillary) - Oil secretion and acne - First seminal discharge

18. A 15-year-old boy is brought to clinic by his parents because of delayed puberty. His growth chart reveals a height just below but parallel to the 5th centile. Examination is unremarkable. Bone age is consistent with a 12 year chronological age. Lab investigations including hormonal study are normal.

Of the following, the **MOST** likely diagnosis is

- A. hypothyroidism
- B. Klinefelter syndrome
- C. hypogonadotropic hypogonadism
- D. craniopharyngioma
- E. constitutional growth delay

Family
Constitutional



19. A 16-year-old boy has delayed puberty; he doesn't develop secondary sexual characteristics; he has a normal stature according to his chronological age. Examination reveals an upper-to-lower segment ratio of 0.8 (normal ratio is more than 0.9).

Of the following, the **MOST** important test to confirm diagnosis is

- A. plasma gonadotropins
- B. growth hormone stimulation test
- C. serum prolactin

3

Hypogonadotropic hypogonadism (HH)
الحصبي مع المنخفضة

• Hypogonadism as a result of gonadotrophin deficiency.

Diagnosis:
قلبي قليل، testosterone، FSH، LH Low
• Small gonads.
• CF. ب. ش. ه. م.

• Absent or delayed puberty.
• Tall stature from delayed closure of epiphyses.
• Normal bone age and age of onset of puberty
• Other family members (often one or both parents) with histories of short stature in childhood, delayed puberty, and eventual normal stature
• Bone age is low for chronological age but normal for bone age

Anosmia and absent olfactory bulbs on MRI in Kallmann's.
MRI = كاشف
الانصباب مائلها يا اما structural مثلًا تيومور او كوشش
functional يعني عنده غزوكة النس مثلاً ريو من لما هو صغيرتي او سكر او مرض نفسي و الي كلها لها علاقة ب body mass index

◆ Hypergonadotrophic hypogonadism Primary hypogonadism
 • testicular failure with normal hypothalamus and pituitary function.
 الخلل ب التمييز
 Diagnosis
 • Low 9 a.m. serum testosterone.
 يرتفعون هنا .
 • Elevated LH and FSH, low inhibin B .
 • causes
 Klinefelter's syndrome (47XXY, male phenotype).
 هذا الكمن كوز
 و هنا اسوي karyotyping علمود الشخصيه
 ما الة علاقة ب الامراي
 Chemotherapy/radiotherapy to gonads.
 Trauma/surgery to gonads.
 Autoimmune gonadal failure.
 Mumps النكاف , Tuberculosis, Haemochromatosis . Anorchidism/cryptorchidism in males.

- D. brain MRI
- E. bone age

20. A 16-year-old boy has delayed puberty and absence of secondary sexual characteristics; he has decreased sense of smell. Examination reveals shortening of the fourth finger. MRI of the brain shows absence of the olfactory bulbs and sulci; abdominal ultrasonography shows absence of the right kidney.

Of the following, the **MOST** likely diagnosis is

- A. Prader-Willi syndrome
- B. Kallmann syndrome
- C. Laurence-Moon-Bardet-Biedl
- D. Klinefelter syndrome
- E. Noonan syndrome

شرح موجود باول مربع

21. A 9-year-old boy develops secondary sexual characteristics manifested by the parents. Examination reveals an enlarged testis of 2.7 cm and presence of pubic and axillary hair. Hormonal assessment reveals increased level of testosterone and gonadotropins.

Of the following, an important **NEXT** step in the management is

- A. abdominal ultrasonography
- B. bone age
- C. bone scan
- D. brain MRI
- E. testicular ultrasonography

Compared with girls, boys with precocious puberty have a higher incidence of CNS disorders, such as tumors and hamartomas, precipitating the precocious puberty.

22. Thyroid disorders consist of a variety of diseases that require different treatment according to the cause.

Of the following, the thyroid disorder that does not require treatment is

- A. congenital hypothyroidism
- B. endemic cretinism
- C. Hashimoto thyroiditis
- D. hyperthyroidism
- E. congenital thyroxine-binding globulin deficiency

It is associated with low serum total T4 concentration, normal TSH, and normal serum free T4. It is a euthyroid condition and does not require treatment with thyroid hormone because it is merely a binding protein abnormality. It is commonly X-linked dominant.

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23. A 16-year-old female adolescent develops swelling at the anterior aspect of the neck with difficulty of the swallowing. Examination reveals a firm, nontender diffuse goiter with a pebble-like surface. Her mother has had a thyroid disease in early adulthood.

Of the following, the **MOST** valuable test to confirm the diagnosis is

- A. thyroid scan
- B. biopsy of the thyroid gland
- C. ultrasonography of the thyroid gland
- D. thyroid function test
- E. serum antithyroid peroxidase

Neither biopsy nor thyroid scan is indicated, although the thyroid scan with reduced uptake may differentiate hashitoxicosis from Graves' disease.

24. An adolescent female develops fever, palpitation and disorientation. Examination shows exophthalmos, goiter, tachycardia and elevated blood pressure. Lab tests reveal elevated T4 and T3 with low TSH.

Of the following, the **BEST** initial treatment is

- A. beta blockers
- B. methimazole
- C. propylthiouracil
- D. radioiodine I^{131}
- E. thyroidectomy

25. A 5-day-old male neonate develops poor feeding, irritability and tachycardia; her mother has been developed hyperthyroidism before pregnancy that was treated by radioiodine. The pregnancy course was unremarkable. Lab investigations reveal markedly elevation of T4 and T3.

Of the following, the **MOST** likely diagnosis is

- A. congenital hyperthyroidism
- B. primary hyperthyroidism
- C. secondary hyperthyroidism
- D. tertiary hyperthyroidism
- E. hyperthyroidism due to radioiodine treatment

وزاري بالبلوبرنت

إذا سال عن most common cause of congenital hypothyroidism

is 1st thyroid Dysgenesis
(2nd) Dysmorphogenesis

26. You are examining a 10-month-old male infant during a routine visit; you find that he has wide wrists. He is still breast fed. X-ray of the wrist joint shows signs of rickets.

Of the following, the **BEST** reflection of vitamin D deficiency is

- A. serum calcium
- B. serum phosphate
- C. measurement of parathyroid hormone
- D. serum concentration of 1,25-hydroxyvitamin D
- E. serum concentration of 25-hydroxyvitamin D

The serum concentration of 25-hydroxyvitamin D is a better reflection of vitamin D sufficiency than the measurement of 1,25-hydroxyvitamin D.

27. A 12-year-old female adolescent is brought to the emergency room because of irritability, paresthesia, and seizure-like state after a psychological trauma. She is conscious with tachypnea and carpopedal spasm.

Of the following, the **MOST** valuable test is the determination of

- A. total serum calcium
- B. ionized calcium
- C. serum pH
- D. serum albumin
- E. serum magnesium

Hypocalcemia

28. Rickets is defined as decreased bone mineralization in growing children. Hypocalcemia may accompany rickets.

Of the following, the condition that causes hypocalcemia rather than rickets is

- A. nutritional vitamin D deficiency
- B. biliary atresia
- C. primary hypoparathyroidism
- D. renal failure
- E. familial hypophosphatemia

29. You are examining a 5-day-old neonate with ambiguous genitalia; there are no palpable gonads in the inguinal or genital area; the blood pressure is normal according to the age.

Of the following, the **NEXT** step in the management of this neonate is

- A. pelvic ultrasonography
- B. chromosomal study
- C. measurement of testosterone level
- D. measurement of the plasma concentration of 17-hydroxyprogesterone
- E. measurement of adrenocorticotrophic hormone (ACTH)

30. A 3-week-old neonate develops repeated vomiting and dehydration. Examination reveals ambiguous genitalia with fusion of the anterior portion of the labioscrotal folds. Ultrasonography reveals presence of uterus, ovaries, and fallopian tubes.

Of the following, the **MOST** valuable test to establish the diagnosis is

- A. plasma concentration of 17-hydroxyprogesterone
- B. aldosterone level
- C. testosterone level
- D. plasma concentration of 11-deoxycortisol
- E. plasma concentration of 18-hydroxycorticosterone

Enzyme deficiency	17-Hydroxyprogesterone	Corticosterone	11-Deoxycorticosterone (DOC)	Sodium	Potassium	Acid-base disorders
21β-hydroxylase	↑↑	↓	↓	↓	↑	Metabolic acidosis
11β-hydroxylase	↑	↓	↑↑	↑	↓	Metabolic alkalosis
17α-hydroxylase	↓	↑↑	↑	↑	↓	Metabolic alkalosis

Enzyme deficiency	Blood pressure	XX (female) genotype	XY (male) genotype
21β-hydroxylase	Hypertension	Female pseudohermaphroditism: clitoromegaly and/or male external genitalia at birth	Normal male external genitalia at birth
11β-hydroxylase	Hypertension	Female pseudohermaphroditism: clitoromegaly and/or male external genitalia at birth	Precocious puberty
17α-hydroxylase	Hypotension	Normal female external genitalia at birth	Precocious puberty

31. A 6-year-old boy develops precocious puberty; he has acne, pubic and axillary hair, and penile enlargement with normal testicular size. His blood pressure is elevated. Lab test reveals hypokalemia, and elevated level of deoxycorticosterone. MRI of the brain is normal.

Of the following, the **MOST** likely enzyme deficiency for this boy is

A. 11-hydroxylase

B. 17-hydroxylase → Delay puberty

C. 18-hydroxylase

D. 18-oxidase

E. 21-hydroxylase → hypotension
→ Metabolic acidosis
↑ Na ↓ K

precocious puberty

32. A 7-year-old boy is going to undergo orthopedic surgery for right total hip replacement; he has been diagnosed with Addison disease 2 years ago and he is on replacement therapy by hydrocortisone since that time.

Of the following, the **MOST** important step in the management of this boy is

A. frequent monitoring of serum electrolytes and glucose

B. start fludrocortisone

C. proceed with surgery

D. increase the maintenance dose of hydrocortisone

E. rigorous use of intravenous fluids

33. Cushing syndrome in children has many causes, either exogenous or endogenous.

Of the following, the **LEAST** likely endogenous cause in children is

A. adrenal adenoma

B. adrenal carcinoma

C. nodular adrenal hyperplasia

D. ACTH-secreting pituitary microadenoma

E. ACTH-secreting tumor

34. An ACTH-secreting pituitary microadenoma resulting in bilateral adrenal hyperplasia is termed Cushing disease; this should be distinguished from Cushing syndrome due to various causes.

Of the following, the **BEST** test that helps to differentiate between Cushing disease and Cushing syndrome is

A. a 24-hour urinary cortisol excretion

B. low-dose dexamethasone suppression test

C. high dose dexamethasone suppression test

- D. late evening salivary cortisol sampling.
- E. serum cortisol level

إن لله عبادة اختصهم بقضاء حوائج الناس، حببهم إلى الخير،
وحبب الخير إليهم، أولئك الآمنون من عذاب الله يوم القيامة"

تم بحمد الله وتوفيقه

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SECTION 23

Endocrinology

ANSWERS

USAMA ALJUMAILY

- 1.(D). Prolactin is the only pituitary hormone that is suppressed by a hypothalamic factor, dopamine. Hypothalamic deficiency leads to a decrease in most pituitary hormone secretions but may lead to an increase in prolactin secretion.
- 2.(E). Six percent of cases of cleft palate are associated with GH deficiency.
- 3.(A). A patient is considered to have impaired fasting glucose, if fasting serum glucose concentration is 100 to 125 mg/dL or impaired glucose tolerance if 2-hour plasma glucose following an OGTT is 140 to 199 mg/dL. Sporadic hyperglycemia can occur in children, usually in the setting of an intercurrent illness.
- 4.(C). Insulin deficiency usually first causes postprandial hyperglycemia and then fasting hyperglycemia. Ketogenesis is a sign of more complete insulin deficiency.
- 5.(A). The presence of polyuria, despite a state of clinical dehydration, indicates osmotic diuresis and differentiates patients with DKA from patients with gastroenteritis or other gastrointestinal disorders.
- 6.(B). The measured serum sodium concentration is artificially low, however, because of hyperglycemia. Hyperlipidemia also contributes to the decrease in measured serum sodium.
- 7.(B).
- 8.(C). Bicarbonate therapy should be avoided as it increases risk of the development of cerebral edema.
- 9.(D). Measurements of HgbA1c are inaccurate in patients with hemoglobinopathies. Glycosylated albumin or fructosamine can be used in these cases.
- 10.(A).
- 11.(E). If ketonuria or ketoacidosis occurs, insulin treatment is necessary to first achieve adequate glycemic control but may be discontinued within weeks with continuation of oral medications.
- 12.(A). This diagnosis should be suspected if extremely high insulin concentrations are detected ($>100/\mu\text{U/mL}$). C-peptide concentrations are low or undetectable, which confirms that the insulin is from an exogenous source.
- 13.(A). The absence of serum and urine ketones at the time of hypoglycemia is an important diagnostic feature, distinguishing hyperinsulinism from defect in counter regulatory hormone secretion. In contrast to hyperinsulinism, serum and urine ketones are positive at the time of hypoglycemia and free fatty acids are elevated.

- 14.(E). Infant has an upper-to-lower ratio of 1.7:1, a 1-year-old has a ratio of 1.4:1, and a 10-year-old has a ratio of 1:1.
- 15.(E). Treatment with GH carries the risk of an increased incidence of slipped capital femoral epiphysis, especially in rapidly growing adolescents.
- 16.(A). In Infants with congenital GH deficiency, the growth rate slows after birth, most noticeably after age 2 to 3 years. These children have an elevated weight-to-height ratio, appearing chubby and short. Male neonates may have a microphallus.
- 17.(C). The normal developmental sequence in girls is thelarche followed closely by pubarche and finally menarche, 2 to 3 years later. In boys the first normal event is scrotal thinning followed by the enlargement of testes and by the appearance of pubic hair.
- 18.(E). Patients with constitutional delay have delayed onset of pubertal development and significant bone age delay. Usually height gain is below, although fairly parallel to, the normal percentiles on the growth curve. The prepubertal nadir, or deceleration before their pubertal growth spurt, is prolonged or protracted. A family history of delayed puberty in a parent or sibling is reassuring.
- 19.(A). Patients with hypogonadotropic hypogonadism have normal proportions and growth throughout childhood and in early puberty. When these patients reach adulthood, eunuchoid proportions may ensue because their long bones grow for longer than normal, producing an upper-to-lower ratio below the lower limit of normal of 0.9 and an arm span greater than their height.
- 20.(B).
- 21.(D). Compared with girls, boys with precocious puberty have a higher incidence of CNS disorders, such as tumors and hamartomas, precipitating the precocious puberty.
- 22.(E). It is associated with low serum total T4 concentration, normal TSH, and normal serum free T4. It is a euthyroid condition and does not require treatment with thyroid hormone because it is merely a binding protein abnormality. It is commonly X-linked dominant.
- 23.(E). Neither biopsy nor thyroid scan is indicated, although the thyroid scan with reduced uptake may differentiate hashitoxicosis from Graves' disease.
- 24.(A). Thyroid storm is a rare medical emergency. Treatment includes reducing the hyperthermia and administering a beta blocker to control the tachycardia, hypertension, and autonomic hyperfunction symptoms.
- 25.(A). Cure of hyperthyroidism before pregnancy (surgery or radioiodine treatment) limits or curtails T4 production but not the underlying immune disturbance producing thyroid stimulating immunoglobulins (TSIs); thus the infant still may be affected, at least transiently.

- 26.(E). The serum concentration of 25-hydroxyvitamin D is a better reflection of vitamin D sufficiency than the measurement of 1,25-hydroxyvitamin D.
- 27.(B). The fraction of ionized calcium is inversely related to plasma pH; alkalosis (resulting from hyperpnea caused by anxiety or from hyperventilation related to physical exertion) can precipitate hypocalcemia by lowering ionized calcium without changing total serum calcium.
- 28.(C). Primary hypoparathyroidism causes hypocalcemia but does not cause rickets.
- 29.(A). The first step toward diagnosis is to determine whether the disorder represents virilization of a genetic female (androgen excess) or underdevelopment of a genetic male (androgen deficiency). Inguinal gonads that are evident on palpation usually are testes and indicate that incomplete development of a male phenotype has occurred. Similarly absence of female internal genitalia (detected by ultrasound) implies that müllerian-inhibiting substance was present and secreted by fetal testes.
- 30.(A). Statistically most virilized females have CAH; 90% of these females have 21-hydroxylase deficiency. The diagnosis is established by measuring the plasma concentration of 17-hydroxyprogesterone and androstenedione, which typically is hundreds of times above the normal range.
- 31.(A). In 11-hydroxylase deficiency, virilization occurs with salt retention, hypokalemia, and hypertension as a result of the excessive production of deoxycorticosterone, a potent mineralocorticoid. Late-onset CAH is typically noted years after birth with pseudo precocious puberty rather than ambiguous genitalia.
- 32.(D). In Addison disease, replacement treatment with 10 to 15 mg/m²/24 hours of hydrocortisone is indicated, with supplementation during stress at three times the maintenance dosage or the use of intramuscular hydrocortisone.
- 33.(E). ACTH-secreting tumor is an extremely rare.
- 34.(C).